
	Division of Environmental Health and Communicable Disease Prevention	
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Creutzfeldt-Jakob Disease (CJD)

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Creutzfeldt-Jakob Disease (CJD)

Overview ^(1,2)

For a more complete description of CJD, refer to the following texts:

- Control of Communicable Diseases Manual (CCDM)
- Red Book, Report of the Committee on Infectious Diseases.

Case Definition ⁽³⁾

1. Sporadic CJD:

Confirmed

- Diagnosed by standard neuropathological techniques, and/or
- Immunocytochemically, and/or
- Western blot confirmed protease-resistant PrP, and/or
- Presence of scrapie-associated fibrils.

Probable:

Progressive dementia and at least two of the following four clinical features:


- Myoclonus – seizure-like severe muscle contractions.
- Visual or cerebellar signs – double or blurred vision, and/or inability to visually recognize familiar objects.
- Pyramidal/extrapyramidal signs – poor control or poor initiation of skilled movement (primarily hands and fingers), poor control of speech, and difficulty forming words.
- Akinetic mutism - no spontaneous movement or attempt to formulate speech.

And

- Atypical EEG during an illness of any duration and/or a positive 14-3-3 CSF assay and a clinical duration to death of less than two years.
- Routine investigations do not suggest an alternative diagnosis.

Suspect:

- Death certificate indicating CJD as a cause of death (this means the mention of CJD anywhere on the death certificate), or
- Progressive dementia and at least two of the following four clinical features:
 - Myoclonus.
 - Visual or cerebellar signs.

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- Pyramidal/extraparidal signs.
- Akinetic mutism.
- And
- No EEG performed or atypical EEG and duration to death of less than two years.

2. Iatrogenic CJD (or Acquired CJD⁽⁴⁾):

- Progressive cerebellar syndrome in a recipient of human cadaveric-derived pituitary hormone, or
- Sporadic CJD with a recognized exposure risk, e.g., antecedent neurosurgery with dura mater implantation.

3. Familial CJD:

Confirmed or probable CJD plus confirmed or probable CJD in a first degree relative, and/or neuropsychiatric disorder plus disease-specific PrP gene mutation.

4. Variant CJD (vCJD) or (nvCJD⁽⁴⁾):

Neuropathological diagnosis is mandatory for confirmation of suspected vCJD.

Confirmatory examination of the brain should show the following neuropathological features:

- Numerous widespread amyloid plaques surrounded by vacuoles.
- Spongiform change most evident in the basal ganglia and thalamus.
- Prion protein accumulation in high density shown by immunocytochemistry, particularly in the cerebellum.

As with sporadic CJD, the patient is suspect for vCJD when the patient's history reveals a rapid dementia and observed loss of muscle coordination. Confirmation of the disease is done through brain biopsy or autopsy.


Information Needed for Investigation

Verify the diagnosis. Obtain copy of the CD-1 or death certificate.

Contact the Regional Communicable Disease Coordinator immediately if vCJD is **suspected**.

Case Follow Up and Control Measures

- **Cases 54 years of age or younger diagnosed with CJD – deceased.** Perform a chart extraction and obtain copies of the following:
 1. Discharge summary

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2. Neurology consultation notes
3. Psychiatric consultation notes
4. EEG reports
5. MRI reports
6. Pathology report from brain biopsy or autopsy

- **Cases 54 years of age or younger diagnosed with CJD – not deceased.** Obtain as much of the above information as is available. If diagnostic laboratory testing has not been performed, inform the physician that laboratory services are available from:

National Prion Disease Pathology Surveillance Center
Institute of Pathology, Room 419
Case Western Reserve University
2085 Adelbert Road
Cleveland, Ohio 44106
Phone: 216-368-0587


The laboratory can perform testing on biopsies of brain tissue, CSF, and blood. Refer to their web site for more information: <http://www.cjdsurveillance.com> (23 May 2003)

- **Cases 55 years of age or older diagnosed with CJD – deceased.** Determine if an autopsy/biopsy or other diagnostic pathology was performed. If so, obtain the pathology report and affix to case report.
- **Cases 55 years of age or older diagnosed with CJD – not deceased.**
 1. Determine if a biopsy or other pathology was performed. If so, obtain the pathology report and affix to case report.
 2. If no laboratory testing has been performed, inform the physician that laboratory services are available for diagnosis from:

National Prion Disease Pathology Surveillance Center
Institute of Pathology, Room 419
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Cleveland, Ohio 44106
Phone: 216-368-0587
Web site: <http://www.cjdsurveillance.com> (23 May 2003)

Control Measures

- See the Communicable Disease Control Manual, Creutzfeldt-Jakob Disease, “Methods of control.”
- See the Red Book, Transmissible Spongiform Encephalopathies, “Control Measures.”

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Laboratory Procedures

For information contact:

National Prion Disease Pathology Surveillance Center
Institute of Pathology, Room 419
Case Western Reserve University
2085 Adelbert Road
Cleveland, Ohio 44106
Phone: 216-368-0587
Web site: <http://www.cjdsurveillance.com> (23 May 2003)


Reporting Requirements

Creutzfeldt-Jakob disease is a Category II disease and shall be reported to the local health authority or to the Missouri Department of Health and Senior Services within three days of first knowledge or suspicion by telephone, facsimile or other rapid communication.

1. For all cases of sporadic CJD, iatrogenic CJD, familial CJD, or variant CJD complete a "Disease Case Report" (CD-1).
2. Contact the Regional Communicable Disease Coordinator immediately for all confirmed, probable, or suspect cases of vCJD.
3. Entry of the complete CD-1 into the MOHSIS database negates the need for the paper CD-1 to be forwarded to the Regional Health Office.
4. All outbreaks or "suspected" outbreaks must be reported as soon as possible (by phone, fax or e-mail) to the Regional Communicable Disease Coordinator. This can be accomplished by completing the Missouri Outbreak Surveillance Report (CD-51).
5. Within 90 days from the conclusion of an outbreak, submit the final outbreak report to the Regional Communicable Disease Coordinator.

References

1. Chin, James, ed. "Creutzfeldt-Jakob disease" Control of Communicable Diseases Manual, 17th ed. Washington, D.C.: American Public Health Association, 2000: 183-186.
2. American Academy of Pediatrics. "Creutzfeldt-Jakob disease". In: Pickering, L, ed. 2000 Red Book: Report of the Committee on Infectious Diseases. 25th ed. Elk Grove Village, IL. 2000: 471-473.
3. Diagnostic Criteria for Creutzfeldt-Jakob Disease (CJD) Global Surveillance, Diagnosis, and Therapy of Human Transmissible Spongiform Encephalopathies: Report of a WHO Consultation, February 9-11, 1998, Geneva, Switzerland.
<http://www.who.int/emc-documents/tse/whoemczdi989c.html> (23 May 2003)

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4. National Prion Disease Pathology Surveillance Center, Case Western Reserve University, Cleveland, Ohio, "Glossary." <http://www.cjdsurveillance.com> (23 May 2003)

Other Sources of Information

Tyler, Kenneth "*Prion Disease* (Prions and Prion Diseases of the Central Nervous System, Transmissible Neurodegenerative Diseases)." Principles and Practice of Infectious Diseases. 5th ed. Eds. Gerald L. Mandell, John E. Bennett, and Raphael Dolin. New York: Churchill Livingstone, 2000: 1971-1985.

Creutzfeldt-Jakob Disease (CJD)

FACT SHEET

What is Creutzfeldt-Jakob Disease (CJD)?

CJD is a rare, fatal brain disorder that causes a rapid, advancing dementia and associated muscle and nerve disturbances. The disease is often referred to as a subacute spongiform encephalopathy because it usually produces microscopic (sponge-like) holes in the brain.

Who gets CJD?

Anyone can be afflicted with this disease. The disease affects both men and women of various ethnic backgrounds usually between the ages of 50-75 years. Typically, CJD onsets in later life and affects 1 person in 1 million people per year worldwide. In the United States there are about 200 cases per year.

How is CJD transmitted/acquired?

There are three general ways through which CJD can be acquired. First, the disease can occur sporadically (at irregular intervals in persons without known risk factors). **Sporadic CJD** is by far the most common type of CJD and accounts for at least 85% of cases in the United States. Second, the disease can be inherited. About 5-10% of cases in the United States are **inherited CJD**. Third, the disease can be acquired through exposure to brain or nervous system tissue/fluids, usually through certain medical procedures. **Iatrogenic CJD** (unintended consequence of a medical procedure) has occurred in cases involving corneal transplants, implantation of electrodes in the brain, dura mater grafts, contaminated surgical instruments, and the injection of natural human growth hormone derived from cadaver pituitaries. In other words, one may become infected with CJD from direct contamination with infected neural tissue/fluid.

CJD is not considered contagious in the traditional sense. Spouses and family members who live with CJD patients have not been found to have a greater risk of getting the disease than the general population.

What are the symptoms of CJD?

Initially, persons may have difficulty sleeping, experience depression, problems with muscular coordination, impaired vision, and personality and behavioral changes such as impaired memory, judgment, and thinking. As the disease progresses, mental impairment becomes severe and involuntary muscle jerks (myoclonus) often occur along with blindness. Eventually, the ability to move or speak is lost and the person enters a coma until death occurs.

How soon do symptoms occur?

Generally, onset of symptoms for sporadic and inherited cases occurs about age 60, can be as early as 20 and as late as 90 years. If acquired by surgical procedures such

as corneal or dura mater transplants, onset of symptoms can be as short as 16 months or as long as 9 years.

How is CJD diagnosed?

A diagnosis of CJD should be considered when an adult develops rapid dementia with loss of muscle coordination. The only way to confirm a diagnosis of CJD is by brain biopsy or autopsy.

How is CJD treated?

Unfortunately, there is no known effective treatment available to cure or control CJD. Current treatment is aimed at controlling symptoms and making the person as comfortable as possible.

What is variant Creutzfeldt-Jakob disease (vCJD)?

vCJD is a rare and fatal human neuro-degenerative condition. It is classified as a transmissible spongiform encephalopathy (TSE) because of characteristic spongy degeneration of brain tissue. vCJD is a new disease, which as of this date, has not been identified in the United States. vCJD is often confused with Creutzfeldt-Jakob (CJD) but reflects different onset and duration characteristics. vCJD is strongly linked to exposure, probably through food, to a TSE of cattle called bovine spongiform encephalopathy (BSE). For more information on vCJD, refer to the Variant Creutzfeldt-Jakob Disease (vCJD) Fact Sheet.

**Missouri Department of Health and Senior Services
Section for Communicable Disease Prevention
Phone: (866) 628-9891 or (573) 751-6113**

Variant Creutzfeldt-Jakob Disease

(vCJD)

FACT SHEET

What is Variant Creutzfeldt-Jakob Disease?

Variant Creutzfeldt-Jakob disease (vCJD) is a rare and fatal human neurodegenerative condition. It is classified as a transmissible spongiform encephalopathy (TSE) because of characteristic spongy degeneration of brain tissue. vCJD is a new disease, which as of this date has not been identified in the United States. vCJD is often confused with Creutzfeldt-Jakob (CJD) but reflects different onset and duration characteristics.

Who gets vCJD?

vCJD affects younger patients (average age 29 years, as opposed to 65 years for Sporadic CJD). It has a relatively longer duration of illness with a median of 14 months (as opposed to 4.5 months for Sporadic CJD). vCJD is strongly linked to exposure, probably through food, to a TSE of cattle called bovine spongiform encephalopathy (BSE). vCJD was first reported in the United Kingdom in 1986.

How is vCJD transmitted/acquired?

The nature of the TSE agent is being investigated and is still a matter of debate. There are several theories under discussion. According to the prion theory, the infective agent is composed largely of a self-replicating protein (prion). Another theory contends that the agent is a "virus-like" agent. The most likely cause of vCJD is exposure to the BSE agent, most plausibly due to dietary contamination by affected bovine central nervous system tissue.

What are the symptoms of vCJD?

Early in the illness, patients usually experience psychiatric symptoms, which most commonly take the form of depression, or a "schizophrenia-like" psychosis. Neurological signs, including unsteadiness, difficulty walking, and involuntary muscle movements develop as the illness progresses. By the time of death, patients become immobile and mute.

How soon do symptoms vCJD occur?

vCJD affects younger patients (average age 29 years) with a relatively long duration of illness (median 14 months).

How is vCJD diagnosed?

Neuropathological diagnosis is mandatory for confirmation of suspected vCJD. Confirmatory examination of the brain should show the following neuropathological features:

- Numerous widespread amyloid plaques surrounded by vacuoles.
- Spongiform change most evident in the basal ganglia and thalamus.

- Prion protein accumulation in high density shown by immunocytochemistry, particularly in the cerebellum.

As with CJD, the patient is suspect for vCJD when the patient's history reveals a rapid dementia and observed loss of muscle coordination. Confirmation of the disease is done through brain biopsy or autopsy.

How is vCJD treated?

There is no known effective treatment available for vCJD. As with CJD, treatment is aimed at controlling symptoms and providing comfort measures.

CJD (Sporadic, Inherited, or Iatrogenic)

For more information on CJD, refer to the Creutzfeldt-Jakob Disease Fact Sheet.

**Missouri Department of Health and Senior Services
Section for Communicable Disease Prevention
Phone: (866) 628-9891 or (573) 751-6113**



MISSOURI DEPARTMENT OF HEALTH AND SENIOR SERVICES
DISEASE CASE REPORT

REPORT TO LOCAL PUBLIC HEALTH AGENCY

1 DATE OF REPORT ____ / ____ / ____		2 DATE RECEIVED BY LOCAL HEALTH AGENCY ____ / ____ / ____	
4 GENDER <input type="checkbox"/> MALE <input type="checkbox"/> FEMALE		5 DATE OF BIRTH ____ / ____ / ____	6 AGE ____
7 HISPANIC <input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> UNKNOWN		10 DATE ARRIVED IN USA ____ / ____ / ____	
8 RACE (CHECK ALL THAT APPLY) <input type="checkbox"/> BLACK <input type="checkbox"/> ASIAN <input type="checkbox"/> PACIFIC ISLANDER <input type="checkbox"/> WHITE <input type="checkbox"/> AMERICAN INDIAN <input type="checkbox"/> UNKNOWN		9 PATIENT'S COUNTRY OF ORIGIN ____	
11 ADDRESS (STREET OR RFD, CITY, STATE, ZIP CODE) ____		12 COUNTY OF RESIDENCE ____	
13 TELEPHONE NUMBER ()		17 DATE OF RETURN ____ / ____ / ____	
14 PREGNANT <input type="checkbox"/> YES (IF YES NUMBER OF WEEKS ____) <input type="checkbox"/> NO <input type="checkbox"/> UNKNOWN		15 PARENT OR GUARDIAN ____	
16 RECENT TRAVEL OUTSIDE OF MISSOURI OR USA <input type="checkbox"/> YES <input type="checkbox"/> NO IF YES, WHERE ____		17 DATE OF RETURN ____ / ____ / ____	

18 OCCUPATION ____		19 SCHOOL/DAY CARE/WORKPLACE ____		ADDRESS (STREET OR RFD, CITY, STATE, ZIP CODE) ____	
20 WORK TELEPHONE NUMBER ()		21 OTHER ASSOCIATED CASES <input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> UNKNOWN IS REPORT PART OF AN OUTBREAK <input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> UNKNOWN		22 TYPE OF COMPLAINT/OUTBREAK <input type="checkbox"/> FOODBORNE <input type="checkbox"/> WATERBORNE <input type="checkbox"/> OTHER (SPECIFY) ____	
23 WAS PATIENT HOSPITALIZED <input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> UNKNOWN		24 PATIENT RESIDE IN NURSING HOME <input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> UNKNOWN		25 PATIENT DIED OF THIS ILLNESS <input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> UNKNOWN	
26 CHECK BELOW IF PATIENT OR MEMBER OF PATIENT'S HOUSEHOLD (HHLD):		PATIENT		HHLD MEMBER	
		YES NO UNK		YES NO UNK	
27 NAME OF HOSPITAL/NURSING HOME ____		IS A FOOD HANDLER			
28 HOSPITAL/NURSING HOME ADDRESS (STREET OR RFD, CITY, STATE, ZIP CODE) ____		ATTENDS OR WORKS AT A CHILD OR ADULT DAY CARE CENTER			
29 REPORTER NAME ____		30 TELEPHONE NUMBER ()		IS A HEALTH CARE WORKER	
31 REPORTER ADDRESS (STREET OR RFD, CITY, STATE, ZIP CODE) ____		32 TYPE OF REPORTER/SUBMITTER <input type="checkbox"/> PHYSICIAN <input type="checkbox"/> OUTPATIENT CLINIC <input type="checkbox"/> PUBLIC HEALTH CLINIC <input type="checkbox"/> HOSPITAL <input type="checkbox"/> LABORATORY <input type="checkbox"/> SCHOOL <input type="checkbox"/> OTHER ____			
33 ATTENDING PHYSICIAN/CLINIC NAME ____		ADDRESS (STREET OR RFD, CITY, STATE, ZIP CODE) ____		34 TELEPHONE NUMBER ()	

35 DISEASE NAME(S) ____	36 ONSET DATE(S) ____ / ____ / ____ ____ / ____ / ____	37 DIAGNOSIS DATE(S) ____ / ____ / ____ ____ / ____ / ____	38 DISEASE STAGE/ RISK FACTOR ____	39 PREVIOUS DISEASE/STAGE ____	40 PREVIOUS DISEASE DATE(S) ____ / ____ / ____ ____ / ____ / ____
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TEST DATE (MO/DAY/YR)	TYPE OF TEST	SPECIMEN TYPE	COLLECTION DATE (MO/DAY/YR)	QUALITATIVE / QUANTITATIVE RESULTS	REFERENCE RANGE	LABORATORY NAME/ADDRESS (INCLUDE STREET OR RFD, CITY, STATE, ZIP CODE)

TREATED (Y/N/UNK)	REASON NOT TREATED	TYPE OF TREATMENT	DRUG	DOSAGE	TREATMENT DATE (MO/DAY/YR)	TREATMENT DURATION (IN DAYS)	PREVIOUS TREATMENT	PREVIOUS LOCATION (LIST CITY, STATE)

SYMPTOM (IF APPLICABLE)	SYMPTOM SITE (IF APPLICABLE)	SYMPTOM ONSET DATE (MO/DAY/YR)	SYMPTOM DURATION (IN DAYS)

44 COMMENTS _____ _____ _____
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41 - DIAGNOSTICS

42 - TREATMENTS

43 - SYMPTOMS

NOTES FOR ALL RELEVANT SECTIONS:

- Stages, risk factors, diagnostics, treatments, and symptoms shown below are examples. To see a more complete listing, please go to <http://www.dhss.state.mo.us/Diseases/DDwelcome.htm>. You may also contact the Office of Surveillance at 1-800-392-0272 for additional information or to report a case.
- All dates should be in Mo/Day/Year (01/01/2001) format.
- All complete addresses should include city, state and zip code.
- Required fields referenced below are italicized and bold, however fill form as complete as possible.

(1) **Date of Report** -- date sent by submitter of document.

(2) Date received will be filled in by receiving agency.

(3-8) **CASE DEMOGRAPHICS/IDENTIFIERS:** *Last name, First Name*, Gender, *Date of Birth*, Hispanic, Race - please check all that apply

(23) Was patient hospitalized due to this illness?

(32) Type of reporter/submitter (doctor, nursing home, hospital, laboratory) (33-34) Attending physician or clinic (full physician name and degree, address, phone)

DISEASE: (35) *Disease name or name(s)*, (36) *Onset date(s)*, (37) *Diagnosis Date(s)*

(38) Disease Stage or Risk Factor**Syphilis**

Primary (chancre present)
Secondary (skin lesions, rash)
Early Latent (asymptomatic < 1 year)
Late Latent (over 1 year duration)
Neurosyphilis
Cardiovascular
Congenital
Other

Gonorrhea or Chlamydia

Asymptomatic
Uncomplicated urogenital (urethritis, cervicitis)
Salpingitis (PID)
Ophthalmia/conjunctivitis
Other (arthritis, skin lesions, etc)

TB Infection

Contact to TB case
Immunocompromised
Abnormal CXR
Foreigner/Immigrant
IV Drug/Alcohol Abuse
Resident, correctional
Employee, correctional
Over 70
Homeless
Diabetes
Healthcare worker
Converter/2 yrs ≥ 10
Converter/2 yrs ≥ 15

(39) *Previous Disease/Stage (if applicable)* (40) *Previous Disease Dates (if applicable)*

(41) Diagnostics (Please Attach Lab Slip)**Test Type****Hepatitis**

Igm Anti-HBc
Anti-HBs
Anti-HBc Total
Igm Anti-HAV
HBsAg
Hep C

TB

Not Done
Mantoux
Multiple puncture device
X-Ray
Smear
Culture

Other

Elisa
Western Blot
Culture
ALT
AST

Specimen Type (blood, urine, CSF, smear, swab), **Collection Date** (Mo/Day/Yr), **Qualitative** (negative, positive, reactive), **Quantitative Results** (1:1, 2.0 mm reading,) **Reference Range** (1:1neg, 1:64 equivocal, 1:128 positive, > 2 positive), **Laboratory** (name, address)

(42) TREATMENT**Reason not treated**

False positive
Previous treated
Age

Drug**TB**

Isoniazid
Ethambutol
Pyrazinamide
Rifampin

(43) SYMPTOMS:

Symptom (jaundice, fever, dark urine, headache) **Symptom Site** (head, liver, lungs, skin), **Symptom Onset Date** (Mo/Day/Yr) and **Symptom Duration** (in days)

(44) **Comments:** Attach additional sheets if more comments needed.